Neurology Case Studies

Second Edition

59 CASE HISTORIES RELATED TO NEUROLOGICAL DISEASES

by

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FOREWORD

This book presents a series of actual case histories drawn from the practices of the authors and ranging over a wide spectrum of neurological disease, both common and esoteric.

In contrast to a CPC approach, in which historical, examination, and laboratory data are given, we have tried to place the reader in the position of the neurological clinician who is caring for the patient, and who must localize the probable site of the disease process, reach a diagnosis, consider differential possibilities, and then choose a treatment plan. The cases presented are interrupted by questions which give the reader the opportunity to make these decisions and then compare his choices and reasoning with those made by the neurologists who were actually managing the cases. In addition, we have asked the reader to consider problems of pathogenic and basic mechanisms underlying the patient's neurological symptoms and signs.

We recognize that experienced clinicians may differ in approaches to the same clinical problem, and the reader is encouraged to pursue his interest in the clinical and basic science questions raised by using the set of references given at the end of each case history. We believe that this "case study" method is both an instructive and enjoyable way to learn clinical neurology.

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NEUROLOGY CASE STUDIES
SECOND EDITION

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CASE STUDY #1

Acute Onset of Double Vision in a Diabetic

HISTORY: A 57 year-old woman, an insulin-dependent diabetic, awoke one morning seeing double. The two images were side by side. The diplopia disappeared when either eye was covered. Visual acuity was 20/20 in each eye.

QUESTIONS:

1. THE SEPARATION OF IMAGES IN THE HORIZONTAL PLANE SUGGESTS THAT THE IMPAIRED EYE MUSCLE IS:
   A. Medial rectus or lateral rectus
   B. Superior rectus or inferior oblique
   C. Inferior rectus or superior oblique

2. IF THE DIPLOPIA PERSISTED AFTER SHE OCCLUDED EITHER EYE, (MONOCULAR DIPLOPIA) THIS WOULD LEAD YOU TO SUSPECT:
   A. Brain tumor
   B. Conversion reaction
   C. Transient ischemic attack
   D. Aneurysm of the posterior inferior cerebellar artery

She reported no headache, eye pain, hearing loss, tinnitus, dysphagia, dysarthria, numbness or weakness of her extremities. She had had a left Bell's palsy one year earlier. This cleared spontaneously three months after onset.

Figure 1

CASE STUDY #1

EXAMINATION: The neurological examination was normal except for decreased perception of vibration at both ankles, and the abnormality of eye movement noted in the photograph on attempted gaze to her left (Fig. 1).

QUESTIONS:

3. THE WEAK MUSCLE IS THE:
   A. Left lateral rectus
   B. Right superior oblique
   C. Left inferior oblique
   D. None of the above

4. THE WEAK MUSCLE IS INNERVATED BY THE:
   A. Oculomotor nerve
   B. Trochlear nerve
   C. Abducens nerve
   D. Optic nerve

5. THE NUCLEUS OF THIS NERVE IS LOCATED IN THE:
   A. Pons
   B. Medulla
   C. Mesencephalon
   D. Diencephalon

YOU WOULD EXPECT THE IMAGE SEPARATION TO BE MAXIMAL AS THIS PATIENT LOOKED:
   A. To her right
   B. To her left
   C. Up and to her right
   D. Down and to her right

7. THE MOST PROBABLE DIAGNOSIS IN THIS PATIENT IS:
   A. Brain stem infarct
   B. Acute diabetic cranial mononeuropathy
   C. Intracranial hemorrhage
   D. Aneurysm of the carotid artery

8. INITIAL TESTS SHOULD INCLUDE:
   A. Tensilon test
   B. 2 hour postprandial blood sugar
   C. Carotid angiogram
   D. Skull X-rays
   E. Pneumoencephalogram

LABORATORY DATA: Complete blood count, VDRL, sedimentation rate, X-rays of chest and skull were all normal. The 2 hour postprandial sugar was elevated. Intravenous injection of 10 mgms of tensilon did not alter the diplopia.

CASE STUDY #1

QUESTIONS:

9. IF THE MUSCLE RESPONSIBLE FOR THE DIPLOPIA IS ONLY SLIGHTLY PARETIC AND CANNOT BE READILY IDENTIFIED, THE RED GLASS TEST IS A USEFUL AID. FOR EXAMPLE, IF, IN THIS PATIENT WITH A WEAK LEFT LATERAL RECTUS, A RED GLASS WERE PLACED IN FRONT OF THE RIGHT EYE, THEN THE RED IMAGE BELONGS TO AND IDENTIFIES THE RIGHT EYE, THE WHITE IMAGE IDENTIFIES THE LEFT EYE. THE PATIENT IS ASKED TO LOOK AT A FLASHLIGHT IN DIFFERENT DIRECTIONS OF GAZE AND TO REPORT THE POSITION WHERE MAXIMUM IMAGE SEPARATION OCCURS. THE EYE THAT LAGS (IN THIS CASE THE LEFT EYE) PROJECTS THE IMAGE FARTHEST IN THE DIRECTION OF GAZE OR, STATED IN ANOTHER WAY, THE MORE PERIPHERAL OF THE DOUBLE IMAGES BELONGS TO THE PARETIC EYE. THUS:
   A. The red and white images would be maximally separated when looking to the left, with the red image then seen to the left of the white
   B. The red and white images would be maximally separated when looking to the right with the red to the left of the white
   C. The red and white images would be maximally separated when looking to the right with the white to the left of the red
   D. The red and white images would be maximally separated when looking to the left with the white to the left of the red

10. THERE ARE MANY NEUROLOGICAL COMPLICATIONS OF DIABETES MELLITUS. EXAMPLES ARE:
   A. Symmetrical motor and sensory polyneuropathy
   B. Polymyositis
   C. Coma
   D. Narcolepsy
   E. Cerebrovascular arteriosclerosis

11. THE MOST LIKELY PATHOGENETIC MECHANISM RESPONSIBLE FOR ACUTE DIABETIC MONONEUROPATHIES, BOTH CRANIAL AND PERIPHERAL, IS:
   A. Degenerative
   B. Ischemic
   C. Nutritional

12. THE MOST COMMON CRANIAL MONONEUROPATHIES OCCURRING IN DIABETICS INVOLVE CRANIAL NERVES:
   A. 1, 3, 5
   B. 2, 4, 7
   C. 3, 4, 10
   D. 3, 6, 12
   E. 3, 6, 7

ANSWERS AND DISCUSSION:

1. A — The medial rectus and lateral rectus muscles are horizontal rotators; the medial rectus is an adductor, the lateral rectus is an abductor. The superior rectus and inferior oblique are elevators, the inferior rectus and superior oblique are depressors. Paresis of any of the latter four muscles will result in vertical diplopia.

CASE STUDY #1

2. B — Some complaints of monocular diplopia are accounted for by early cataracts, uncorrected irregular astigmatism or subluxated lenses. Rarely it may be cerebral in origin. Most commonly, the complaint indicates hysteria or malingering.

3. A — Note that the photograph indicates a total inability of the patient to abduct her left eye.

4. C

5. A

6. B — Separation of images is greatest on gaze to her left, in the field of action of the paretic left lateral rectus muscle.

7. B — The brain stem is compact, with many important structures confined to a small area. When infarction occurs at this site, isolated injury to a single cranial nerve is not likely; rather, there may be various combinations of injury to cranial nerve nuclei, to long motor and sensory tracts and to cerebellar connections.

Intracranial hemorrhage is usually characterized by severe headache, which may be followed by convulsions, hemiparesis or a depressed level of consciousness. Blood in the subarachnoid space may produce signs of meningeal irritation: nuchal rigidity and a positive Kernig's sign.

An aneurysm of the internal carotid artery is a common cause of an isolated third nerve palsy. Rarely, an infraclinoid aneurysm may give rise to an isolated sixth nerve palsy.

8. ABD — All patients with diplopia in whom the diagnosis is not readily apparent deserve a tensilon test to rule out myasthenia gravis. Skull films may reveal bony erosion, hyperostosis, abnormal calcification or pineal shift, thus providing important clues to diagnosis. Intracranial contrast studies are never indicated as initial studies.

9. D

10. ACE — Acute cranial and peripheral mononeuropathies as well as symmetrical polyneuropathy occur in patients with diabetes mellitus. Coma can occur associated with acidosis and ketosis, or with marked hyperglycemia but without ketoacidosis (hyperosmolar nonketotic coma). Coma may result also in diabetics who have taken excessive medication and have developed hypoglycemia.

11. B — Walsh and Hoyt have stated that ischemia from occlusive changes in small vessels is the essential cause of acute diabetic mononeuropathies. Raff[3] has suggested that diabetic asymmetric mononeuropathies may be vascular in origin whereas symmetrical polyneuropathies may be metabolic in origin. Dreyfus et al.[4] described a diabetic who developed a sudden third nerve palsy, and died five weeks later. Pathologic examination suggested an incomplete ischemic neuropathy. Weber et al.[6] suggested that in a typical diabetic third nerve palsy, "the complete or relative preservation of pupillary reaction, a valued sign in the recognition of this disorder, is apparently the result of sparing of the peripheral portion of the nerve, where pupillomotor fibers are said to traverse."

12. E

CASE STUDY #1

FOLLOW UP: This patient was reexamined two months later. She no longer complained of diplopia, and there was now a full range of ocular motion.

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CASE STUDY #2

Weakness in a Young Boy

HISTORY: The illustration is that of the patient at age seven (Figs. 2A and B). His prenatal history is unremarkable. He was a full-term, uncomplicated delivery with no evidence of neonatal distress. He sat at seven months of age and walked at eighteen months. In retrospect, his parents stated that he always seemed to have a peculiar gait but could be no more specific than this. At age four, his family became increasingly concerned when he began to fall easily, arose somewhat slowly, and was noted to have difficulty in climbing stairs. The family pediatrician, after checking the child over, was unable to find any definite abnormality and suggested a period of further observation.

QUESTION:

1. TRUE OR FALSE:
   A. Abnormality of gait is a common hysterical symptom in children
   B. In a myopathy, proximal weakness is more common than distal
   C. In a neuropathy, proximal weakness is more common than distal
   D. Pain is an uncommon feature of childhood neuromuscular disease
   E. In primary muscle disease the sensory pathways are usually spared

At age five, progressive gait difficulty was clearly present, he began to fall more frequently, and he was unable to hold heavy objects. His school playmates took advantage of his problem, frequently pushing him over and making fun of him during play. His mother noted that his back was "swayed" and his abdomen prominent.

CASE STUDY #2

Figure 2A

Figure 2B

CASE STUDY #2

A maternal uncle had developed muscular weakness at age seven which was progressive in nature. This uncle became wheelchair-confined in his early teens and expired from respiratory insufficiency and progressive debilitation at age twenty. The patient has one sister, age nine, who is in good health. Otherwise there is no family history of neuromuscular disorder or other significant illnesses.

QUESTION:

2. THIS FAMILIAL PATTERN OF DISEASE IS MOST SUGGESTIVE OF A TYPE OF INHERITANCE TERMED:
   A. Autosomal dominant
   B. Sex-linked recessive
   C. Autosomal recessive
   D. Sporadic
   E. None of above

EXAMINATION: Blood pressure 105/65 right arm, sitting. Pulse 84 and regular. The heart was not enlarged. The lungs were clear and no organomegaly was present. Increased lumbar lordosis was noted as well as prominent scapular winging and protuberance of the abdomen. There was suggestive enlargement of the calves bilaterally. He was unable to walk on his heels because of tibialis anterior weakness and contracture of the heel cords. When arising from a supine position, the use of his upper extremities was required to elevate the trunk. There was questionable minimal facial weakness bilaterally. There was moderate weakness of the proximal extremity muscles of all four limbs in a symmetrical manner. The neck flexors were weak. All deep tendon reflexes were absent. There was no cerebellar deficit. The sensory examination was intact to all modalities. No fasciculations were noted and there was no myotonia.

QUESTIONS:

3. THE DESCRIPTION OF HIS LOWER EXTREMITIES SUGGESTS:
   A. Cerebellar deficit
   B. Spastic paraparesis
   C. Pseudohypertrophy
   D. Peripheral neuropathy
   E. Denervation

4. THE EPONYM APPLIED TO HIS DIFFICULTY IN RISING IS:
   A. Gower's sign
   B. Babinski's sign
   C. Brudzinski's sign
   D. Charcot's triad
   E. Myerson's sign

LABORATORY DATA: CBC, urinalyses and routine blood studies were all within normal limits. Serum SGOT 185, SGPT 160, Aldolase 102, CPK 500. Electrocardiogram was within normal limits as was an electroencephalogram. Electromyography showed polyphasic "miniature" motor units of markedly reduced voltage and duration. Rare fibrillatory activity was observed in the quadriceps on the right. These abnormal motor units were found mainly in the clinically affected muscles but in apparently normally strong muscles as well. Right median nerve conduction time was normal as was repetitive stimulation of the median nerve. A right quadriceps muscle biopsy was carried out. This revealed moderate variation of fiber size with scattered focal muscle necrosis and phagocytosis. There was a moderate increase of connective tissue and fat. The blood vessels and nerve terminals observed were normal.

QUESTIONS:

5. THE MOST LIKELY SITE OF THE DISEASE IS:
   A. Spinal cord
   B. Anterior horn cell
   C. Peripheral nerve
   D. Neuromuscular junction
   E. Muscle fiber

6. THE MOST LIKELY DIAGNOSIS IS:
   A. Duchenne muscular dystrophy
   B. Myasthenia gravis
   C. Guillain-Barré syndrome
   D. Polymyositis
   E. Friedrich's ataxia

CASE STUDY #2

7. INDICATE WHICH OF THE FOLLOWING ARE CONSIDERED AS TYPES OF MUSCULAR DYSTROPHY:
   A. Duchenne
   B. Polymyositis
   C. Limb-girdle
   D. Facioscapulohumeral
   E. Charcot-Marie-Tooth

8. THE MOST SENSITIVE INDICATOR OF A MYOPATHIC DISORDER IS:
   A. Urinary creatine
   B. Urinary creatinine
   C. Serum creatine phosphokinase (CPK)
   D. Serum creatinine
   E. Serum lactic acid

ANSWERS AND DISCUSSION:

1. A. False; B. True; C. False; D. True; E. True

   In almost all myopathies, with the exception of myotonic muscular dystrophy and a few very rare conditions, the preponderance of muscle wasting and weakness is proximal. This is in contradistinction to neuropathic disorders, such as diabetic neuropathy where the atrophy and weakness is more pronounced in the distal extremities. If weakness and wasting is present without sensory involvement, this suggests that the disease is located within the motor unit, i.e., from the anterior horn cell on out to the muscle fiber itself. Although purely motor neuropathies do occur, these are very rare.

2. B The hallmark of autosomal dominant inheritance is the occurrence of the disorder in both parent and child. In autosomal recessive inheritance, involvement of one generation only is the rule. The term sex-linked recessive refers to the fact that the abnormal gene or genetic defect is located on the sex, usually X, chromosome. In this situation, the female becomes the carrier while only males are affected.

3. C Pseudohypertrophy is a phenomenon limited almost entirely to muscular dystrophies. It refers to muscle tissue which becomes enlarged, but is in fact weak, which differentiates it from physiologic or work hypertrophy. This occurs as normal functioning muscle tissue is progressively replaced by fat and connective tissue. In Duchenne Muscular Dystrophy, which is the condition where pseudohypertrophy is most frequent, it is usually most prominent in the calves and thighs, but occasionally may be quite generalized, giving rise to an "infantile Hercules" appearance.

4. A Gower's sign refers to a phenomenon of the child "walking up" his lower extremities with his arms to achieve the upright position. This occurs when there is weakness of the pelvic musculature and low back muscles. It is not specific for muscular dystrophy, but can occur in any situation where weakness in these areas is present.

5. E

6. A